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NIH Fiche to Paper Journal

TITLE:	JOURNAL OF CRANIOFACIAL SURGERY
PUBLISHER/PLACE:	Little, Brown And Company Boston Ma
VOLUME/ISSUE/PAGES:	1998 Sep;9(5):433-7 433-7
DATE:	1998
AUTHOR OF ARTICLE:	Cascone P; Valentini V; Nicolai G; Agrillo A
TITLE OF ARTICLE:	A rare complication of craniofacial fibrous dyspla
ISSN:	1049-2275
OTHER NOS/LETTERS:	Library does NOT report holding title
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A Rare Complication of Craniofacial Fibrous Dysplasia

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Fibrous dysplasia is a rare disease, the cause of which in not entirely known. The most common clinical manifestations of fibrous dysplasia of the temporal bone are the presence of two different pathologic disorders: morphologic and functional. In this study, a rare complication of fibrous dysplasia with temporal localization is examined, and includes a description of a clinical case considered by the authors to be emblematic both for the rarity of the area involved and for the peculiarity of the initial manifestations.

Key Words: Fibrous dysplasia, temporal bone

Fibrous dysplasia is a multiform chronic metabolic osteopathy, because of both its clinical manifestations and the course of its development.[1] It is characterized by alterations in the normal bone tissue structure that result in complete growth failure and progressive replacement with fibrous tissue rich in variably combined collagen, fibroblasts, and osteoid tissue.[2]

Different clinical forms of fibrous dysplasia can be divided into two categories. The first is a form with an exclusively osseous localization, also called Jaffe-Lichtenstein disease. This localization can be unifocal (also called the monostotic form), occurs 70% of cases, and effects, with decreasing frequency, the ribs, the femur, the maxilla, the jaw, and, less frequently, the other bones of the cranium, the phalanxes, and the vertebral column). The plurifocal form (also called polyostotic form), occurs 30% of cases, and effects mainly the lower half of the body, although in 40% to 50% of cases it is located at a craniofacial level.[1,3–5] The second is a form in which there is a combination of plurifocal fibrous dysplasia, cutaneous phenomena (such as hyperchromia), and endocrine disorders such as precocious puberty in girls, hyperthyroidism, hyperparathyroidism, Cushing disease, and diabetes insipidus. This second form is also known as McCune-Albright syndrome.[1,3–5] The monostotic form is twice as frequent as the polyostotic form and approximately 30 times more frequent as McCune-Albright syndrome.[6] In the monostotic form, the incidence of craniofacial localization is about 10%, whereas in the polyostotic form, with widespread skeletal disseminations, the incidence of craniofacial localization is 100%.[7,8] In the polyostotic form the temporal bone is involved in 18% of the cases.[7,8]

Fibrous dysplasia is a rare disease, representing 2.5% of all bone tumors and 7% of benign bone tumors.[1,4,5,9–11] It affects women more often than men (2:1 ratio) and it is generally diagnosed during infancy or adolescence.[11,12] The cause of fibrous dysplasia is debatable and partially unknown.[1,3,4,10,13,14] The proposed causes of dysplasia reported in literature are: abnormal enzymatic activity involved in mesenchymal bone formation,[15] disorders in metabolism of calcium and phosphorus, and hyperplasia of the osteoblasts.[3] Although fibrous dysplasia is usually considered a histologically benign lesion, it presents a stated risk of sarcomatous transformation (0.4–4%).[4,7,11,16] It is also reported[16] that the incidence of malign degeneration is 400 times greater in patients who undergo radiation therapy compared with those who do not. The diagnosis of fibrous dysplasia is made not only by clinical observations, but also on the basis of radiologic and histologic exams.[17]

The maxillofacial localizations, can be classified as follows.[1,11] Ophthalmologic localizations include exophthalmus, ambiopia, visual field disturbances, blindness (as a consequence of compression of the eyeball, the optic nerve, and the oculomotor nerve), and epiphora (as a consequence of compression of

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the lacrimal canal).[1,11] Oronasalsinusal localizations include sinusitis, nasal obstruction, epistaxis, anosmia, dental eruption disorders, and dental mobility and displacement.[1,11] Neurologic localizations include cephalalgia, facialalgia, and neuropsychic disorders.[1,11] Extrabony clinical signs can be found in conjunction with the above, for instance: pigmentary and endocrine disorders (the latter represented by precocious puberty in girls, hyperthyroidism, hyperparathyroidism, Cushing disease, gynecomastia, diabetes insipidus).[1,7,11] Furthermore, a cholesteatoma of the external auditory conduct is found frequently as a result of compressive stenosis.[11,18]

Fibrous dysplasia can have three types of radiologic images.[1,5,11,18] The pagetoid type (found in 39%–56% of cases) is a mixed form having both radiotransparent and radiopaque images. This type is frequently observed in craniofacial localizations. The sclerotic type (found in 23%–27% of cases) is the compact form, characterized by a homogeneously radiopaque image. It is frequently observed in the facial eminence. The pseudocystic type (found in 21%–34% of cases) appears with osseous rarefaction images in a spherical or oval shape. This is the most common in the jaw, with a radiologic aspect called *soap bubble* or *bee-hive*.[1,5,11,18,]

The bones interested show macroscopically more or less extended and complete substitution of the bone tissue with a whitish or reddish tissue, the result of intense vascularization.[1,11,19–21] The microscopic exam reveals that this tissue is made of a rather flail fibrillar connective tissue, disposed in variously orientated bundles, with an occasionally whirling structure. Within this tissue there are osseous trabeculae of different sizes that originate from metaplasia of the fibrous tissue and therefore are not surrounded by osteoblasts (this aspect distinguishes this tissue from the ossifying fibroma in which the osseous trabeculae are, on the contrary, surrounded by the osteoblasts). Pseudocystic structures originating from a degenerative process and little hemorrhagic centers surrounded by giant plurinuclear cells can also be present.

Fibrous dysplasia differs from cherubism, in which the excessive growth of fibrous tissue exclusively interests the jaw. Furthermore, in the fibrous tissue of cherubism, the ossifying processes are not observed.[1,11,19,20,21]

In this study, a rare complication of fibrous dysplasia with temporal localization is examined, beginning with a description of a clinical case we consider emblematic both for the rarity of the area involved and for the peculiarity of the initial manifestations.

Case Report

In June 1996, we observed a 38-year-old ma, C.V., with reduced function of the temporomandibular joint manifested by a limited ability to open his mouth and a joint crepitus in the right temporomandibular joint present while opening his mouth. The patient's history revealed that in June 1988, in another medical center, he underwent plastic surgery of the right external auditory conduct for the presence of an osteoma.

External objective examination of the maxillofacial region, in the prospective and lateral views, revealed a tumescence in the right parotid, masseteric, and submandibular regions. This tumefaction, exceeding 3 cm, was neither sore nor painful to touch.

Clinical examination revealed a partial stenosis of the external auditory conduct. The mandibular function tests showed limitations of the maximum aperture and lateral movements on the left, with the maximal opening of 26 mm. The palpation and auscultation revealed a clicking sound from the right temporomandibular joint while opening and closing.

In August 1996 the patient underwent an electrognathographic examination (Fig 1) that revealed disorders in orientation, distribution, and kinetics of the traces on all data planes. In particular, there was a reduction of the values of maximum opening (28.4 mm) with lateral deviation toward the right during the intermedioterminal phase of the opening and a general reduction of the absolute speed values during all phases of opening and closing. During the mandibular lateral excursion movements, both during maximum intercusping and disimpaction, there were limitations in the left lateral movements (2.7 mm). Furthermore, the exam recorded limited protrusion movement (5.0 mm), and the masticatory and occlusal function tests revealed vertical morsel cycles shifted toward the right. The global electrognathographic examination showed alterations in mandibular function with gnathographic irregularities consistent with a right articular block.

In the same month, the patient was underwent a computed tomography scan of the facial eminence (Fig 2) that revealed the presence of a newly made tissue with osseous density situated in the right retromandibular region. This mass extended craniocaudally for approximately 4 cm with a pouring aspect involving and adhering to part of the homolateral temporal bone, and with extensive involvement of the glenoid cavity and minimal involvement of the ala major ossis sphenoidalis. These structures showed signs of osseous adjustments and thickening

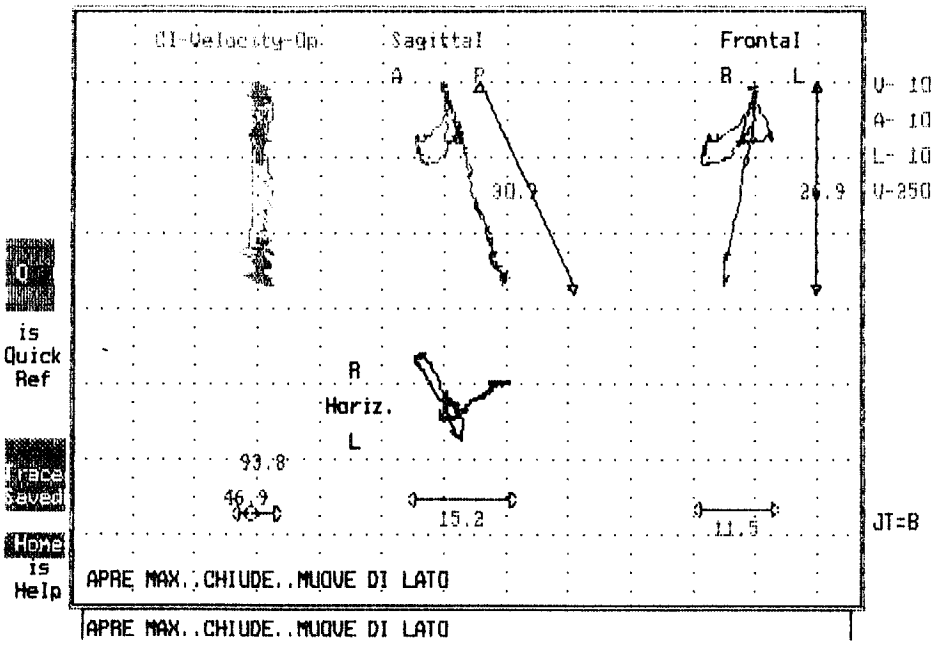

Fig 1 Preoperative electrognathographic exam. Note the reduction in values of maximum opening of the mouth associated with lateral deviation toward the right. Limitation of the left lateral and protrusion movements are present.

of the internal trabeculae, presumably indicating an ossifying process of a fibrodysplasic nature.

In January 1997 the patient underwent surgery that was performed by making a right preauricular, pretragal, and temporoextended skin incision, and ablating the portion of retromandibular dysplastic tissue, which had supported the development of a cholesteatoma in the anterior portion of the external auditory conduct. An arthroscopy of the upper articular compartment of the right temporomandibular joint was performed and revealed hyperemic edema of the bilaminar area, hyperlucent areas that deformed the normal profile of the glenoid tectum and the medial wall, and the presence of microvillus processes in the posterior area of the upper compartment. No abnormality was present in the disc. The zygomatic tubercle, the glenoid cavity, and the tympanic bone were remodelled to improve articular function.

The ablated material was sent for histopathologic examination which confirmed the diagnosis of fibrous dysplasia of the temporal bone. The postoperative period passed without significant complications. During this period, functional therapy of the temporomandibular joint was carried out (Fig 3).

In November 1997 the patient underwent an electrognathographic exam (Fig 4) that revealed a re-

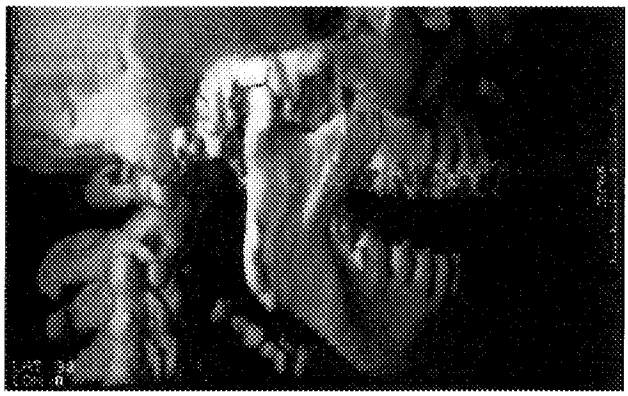

Fig 2 Preoperative three-dimensional computed tomography scan of the facial eminence. The exam shows the presence of a newly made tissue with osseous density situated in the retromandibular region with a "pouring" aspect that involves part of the temporal bone and glenoid cavity, and minimally involves the ala major ossis sphenoidalis.

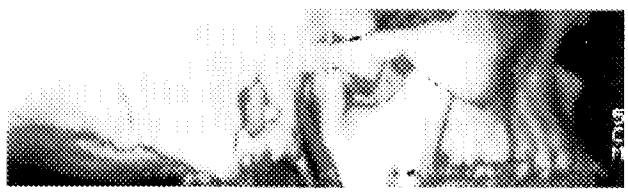

Fig 3 Postoperative three-dimensional computed tomography scan of the facial eminence. The exam shows significant variations with respect to the previous exam. Note the remodelling of the zygomatic tubercle and the glenoid cavity.

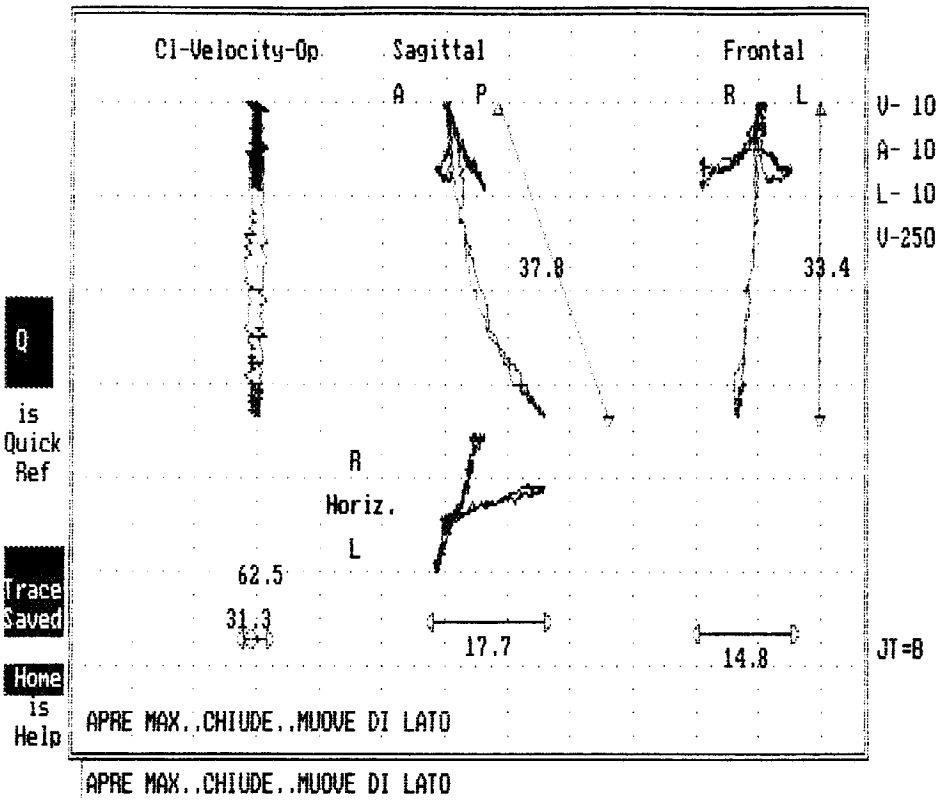

Fig 4 Postoperative electrognathographic exam. The exam shows improvement in articular function during movements of maximum aperture, lateral movements, and protrusion.

markable improvement in the mandibular movements, including increased values of maximum aperture (37.1 mm), right and left lateral excursion (9.4 mm and 5.4 mm, respectively), and protrusion (8.5 mm). Regarding the preoperative exam of August 1996, there remained a diffuse reduction of the absolute speed values during all phases of opening and closing and a mild lateral deviation toward the right during the intermedioterminal phase of opening.

Discussion and Conclusions

The most common clinical manifestations of fibrous dysplasia of the temporal bone are the presence of two pathologic disorders: morphologic and functional.[3,7,9–11,18,] The morphologic disorders are usually caused by the presence of a progressive tumefaction that is hard and not painful, and determines distortion and asymmetry of the temporomandibular area.[9]

The lesion may induce progressive stenosis of the external auditory canal followed by gradual reduction of hearing and possibly complete hearing loss.[7,10,11,18] Furthermore, functional disorders associated with pain resulting from compression of cranial nerves and nearby anatomical structures may be detected.[1,4,7,11] The development of a cholesteatoma can frequently be detected as a consequence of the stenosis of the external auditory conduct.[11,18] In initial cases, the lesion may be diagnosed only after radiographs are taken for other reasons.[11]

As in our patient, fibrous dysplasia may manifest initially as general joint disturbances. However, when he underwent temporomandibular joint function tests, a limited ability to open the mouth and make other mandibular movements was revealed and observed by an electrognathographic exam.[22]

Radiologically, the differential diagnosis of fibrous dysplasia of the temporal bone should be made with ossifying fibroma, cysts, Paget osseous disease, osteochondroma, giant cell granuloma, osteoma, and osseous metastasis.[3] Computed tomography scanning is useful in diagnosis and necessary to define the extension of the lesion toward the basis cranii and the cerebral matter (and the anatomic relationship with nearby structures). Dynamic scintigraphy is necessary to highlight the mono- and polyostotic aspects of fibrous dysplasia.[11]

We believe that the electrognathographic exam plays an important role in identifying fibrous dysplasia of the temporal bone, especially when it effects the homolateral temporomandibular joint followed

by articular limitations. Many pathologic conditions, in fact, limit one's ability to open one's mouth, such as lock, temporomandibular joint ankylosis, hypercondolism, chondromatosis, and muscular problems. We therefore believe that the main symptom of temporal localization of fibrous dysplasia may be a limitation in the ability to open the mouth and suggest that to make a correct differential diagnosis, a morphofunctional examination of the temporomandibular region be conducted. It is possible to perform an objective study of mandibular function with the aid of an electrogastrography exam by quantifying precisely the seriousness of the disfunction and by recording the induced changes from the beginning to the end of the therapeutic treatment.[22] As reported in our patient, this method allowed us to measure articular function before and after surgery.

The surgical treatment of fibrous dysplasia is still debated among authors.[1,7,9,11,13] Not long ago the most common surgical technique was to reshape the osseous district effected,[2] whereas now, with the help of the latest radiologic techniques, it is possible to outline the margins of the dysplasia with greater accuracy by analyzing the computed tomography images. This also makes radical ablation possible with favorable anatomic structure. Furthermore, the improvement of rebuilding techniques through eterologous grafts or autologous-free (or revascularized) bone has allowed the immediate reconstruction with good morphofunctional results. However, the remodelling technique can still be applied to anatomic districts that are difficult either to approach or remove, such as the structures of the skull base (as in out patient). We believe that a remodelling technique should be performed rather than radical ablation, particularly in patients of developmental age in whom it is thought necessary to undertake periodic controls at least until the end of growth.[2]

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